

Posters

11. Gastroenterology/Liver

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[245] A 10-year retrospective study of cystic fibrosis patients with distal intestinal obstruction syndrome (DIOS)

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Objectives: To describe our experience of DIOS management in an adult CF centre over the past 10 years.

Methods: We assessed all cases of DIOS between 2002 and 2012 as captured by electronic patient records and UK registry data. We reviewed imaging to ensure episodes of DIOS met strict criteria.

Results: 37 patients (mean age 35 yrs, SD 12.0; 21 male) had 47 episodes of DIOS, an annual incidence of 0.85%. 56% (19/34) were DF508 homozygous and 35% (12/34) were DF508 heterozygous. All patients were on pancreatic enzymes. Mean FEV1 was 56% (SD 31.0) and mean FVC was 74% (SD 27.2). Abdominal CT was performed on 13 patients. Treatments included oral gastrografin (88%), "Kleen-Prep" (56%), macrogols (56%), N-acetyl cysteine (18%), phosphate and gastrografin enemas (26% and 12%). 62% received intravenous fluids and 32% a nasogastric tube. Colonoscopy with gastrografin instillation was performed for 6 patients (13%). 1 patient required 2 colonoscopies, then was treated with neostigmine with excellent effect. 2 patients presented at other hospitals and underwent surgery. On discharge 89% received macrogol laxatives and 37% oral gastrografin. 1 patient was discharged on prucalopride with good effect. No patients died during their admission. Comparison with 37 age and sex matched CF controls showed more DIOS patients were pancreatic insufficient (37 vs 27, $p < 0.001$) and had a history of meconium ileus (12 vs 2, $p < 0.01$).

Conclusions: Our study shows the relatively low incidence of DIOS in adults. Most cases were managed conservatively but 13% of patients required gastroenterology input for colonoscopic therapy. Neostigmine and prucalopride may be considered in highly selected cases.

[247] Diagnoses of cystic fibrosis liver disease in an adult cystic fibrosis centre

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Objectives: Estimates of the prevalence of Cystic Fibrosis Liver Disease (CFLD) vary, diagnosis commonly being made in adolescence. Diagnosis is based on physical examination, persistent abnormality of liver function tests (LFTs), and ultrasound scan (USS) abnormality. The role of ultrasound to detect early CFLD is questioned, as is the use of ursodeoxycholic acid (UDCA). We reviewed diagnoses of CFLD in our adult CF centre.

Methods: Retrospective review of patient notes and electronic records (from previous five years) of 126 people over 17 years who attend Bristol Adult CF Centre. 87% of patients had abnormal LFTs, with 27% having persistent abnormalities. Rises in transaminases and alkaline phosphatase were equally common with hyperbilirubinaemia seen in <15% patients.

31% had an abnormal USS, 65% of these showing fatty liver. Of 43 patients identified as having "known" CFLD, 20 had a normal liver USS and 12 had parenchymal disease or overt cirrhotic change. Splenomegaly was seen in only 3 patients. 10% did not attend for USS. 11% of those with normal LFTs had an abnormal USS, and 71% with abnormal LFTs had a normal USS.

UDCA doses ranged from 250 mg to 750 mg, with no correlation between dose and weight.

9 patients underwent FibroscanTM liver elastography. 15 patients had endoscopy (for various indications), 3 being found to have oesophageal varices.

Conclusion: This review confirmed previous findings of poor correlation between liver function, USS abnormalities, existing diagnoses of "CF liver disease", OGD or elastography findings and dosing of UDCA. We plan to investigate the role of liver elastography in rationalising investigations in CF liver disease.

[246] High frequency of liver abnormalities in adult cystic fibrosis patients after lung transplantation

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Background: CF associated liver disease (CFLD) mainly develops in early childhood and occurs in up to 30% of CF patients including features of steatosis, fibrosis and cholestasis. We aimed at characterizing liver disease in adult patients with CF after lung transplantation (LuTx).

Methods: Consecutive patients admitted to our outpatient clinic received ultrasound, fibroscan, MRI/MRCP as well as testing of liver function tests, lipids and vitamin D. Liver diseases of other origins were excluded.

Results: 36 patients (median age 30 y, IQR 25–38 y; 42% male) were included in the study. Median time since LuTx was 4.5 y (IQR: 2.3–8.3 y). Elevated AP/GGT were found in 33%, elevated ALT/AST levels in 28% of patients. In 43% (12/23) of patients steatosis and in 18% (4/23) either fibrosis or liver cirrhosis was diagnosed by ultrasound. In 39% (7/18) of patients MRI/MRCP revealed cholangiectasia, and delayed excretion of contrast medium in 20% (3/15). Fibroscan proved cirrhosis in 2 patients, whereas in all other patients fibrosis 0 to 1 was detected. No association of liver function tests and results of imaging was found. Steatosis was significantly associated with hypertriglyceridemia ($p < 0.05$), but not with hypercholesterolemia. Vitamin D levels in general were low. However, neither an association of vitamin D, nor gender or BMI with liver disease was evident.

Conclusion: About 40% of post transplant patients show bile duct alterations suggesting presence of CFLD. Hepatic steatosis associated with hypertriglyceridemia is a common finding in adult CF patients after LuTx. Liver function tests seem to be a weak parameter for monitoring liver disease in CF patients.

[248] Evaluation of the clinical picture, considering liver lesions in infants with cystic fibrosis diagnosed during newborn screening

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The aim of the study was to evaluate the relationship between the genotype, clinical picture and features of liver damage in infants with cystic fibrosis (CF) diagnosed during newborn screening.

Patients and Methods: The analysis involved 42 children, aged from 2 to 6 months, in whom CF was diagnosed on the basis of screening tests. The analysis included: genotype, birth weight, Apgar score, clinical symptoms, parameters of pancreatic exocrine function, markers of liver damage and cholestasis, and abdominal ultrasound abnormalities. The obtained results were statistically analysed.

Results: The most common mutations of CFTR gene observed in infants with diagnosed CF were: mutation delF508 – 36/42 – 85.7% [homozygotes delF508/delF508 – 13/42 – 30.9% – heterozygotes delF508/other mutations 23/42 – 54.7%) and 1717–1G/A/2143delT in 2/42. The clinical picture was dominated by respiratory symptoms (16/42) and/or body mass deficiency (9/42). In 24/42 infants elevated acid steatorrhea in faeces was detected. No statistically significant differences were observed between clinical symptoms, the birth weight, the birth Apgar score and mutation of CFTR gene. Increased aminotransferase activity was observed in 10/42 infants, whereas GGTP in 15/42. In 7/42 children the abdominal ultrasound revealed hepatic abnormalities, most often in the form of liver enlargement and/or increased echogenicity.

Conclusion: Features of liver damage and/or cholestasis were observed even in the youngest patients with diagnosed cystic fibrosis. Mutation delF508 was the most common abnormality of the genotype, which could have some impact on the manifestation of hepatic abnormalities.